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Clinical Assessment of Chorea Severity

The hallmark presentation of Huntington's disease (HD) includes motor, cognitive, and psychiatric symptoms.¹ Chorea is the most visible of the motor symptoms and affects 90% of HD patients at some point during their illness.²

Chorea is typically assessed as part of the Unified Huntington's Disease Rating Scale (UHDRS)—a validated and reliable research tool developed by the Huntington Study Group (HSG).³

Xenazine does not cure the cause of HD chorea and does not treat the other symptoms of HD.

UHDRS* Total Chorea Score¹

Assessing Maximal Chorea

- 0 = absent
- 1 = slight/intermittent
- 2 = mild/common or moderate/intermittent
- 3 = moderate/common
- 4 = marked/prolonged

*UHDRS is a validated and reliable research tool developed by the HSG to provide uniform assessment of the clinical features and course of Huntington's disease (HD). Chorea for each body part is scored from 0 to 4 and the score ranges from 0 to 28.

Body Parts Scored

- Face ☐
- Bucco-oral-lingual ☐
- Trunk ☐
- Right upper extremity ☐
- Left upper extremity ☐
- Right lower extremity ☐
- Left lower extremity ☐

HD Patient Videos[†]

Patient Case 1: Leslie

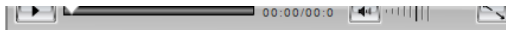
Patient presenting with a pre-treatment total chorea score (TCS) of 15 and a TCS of 7 at 10 weeks post-treatment



Patient Case 2: Gary

Patient presenting with pre-treatment total chorea score (TCS) of 9 and a TCS of 7 at 18 weeks post-treatment





*These are videos of actual patients being evaluated by their physician. Individual results may vary. Xenazine may not be effective in reducing choreic movements in all HD patients. See Important Safety Information below.

[Xenazine Clinical Trial Data »](#)

Indications and Usage:

Xenazine is indicated for the treatment of chorea associated with Huntington's disease.

Important Safety Information:

DEPRESSION AND SUICIDALITY

Xenazine can increase the risk of depression and suicidal thoughts and behavior (suicidality) in patients with Huntington's disease. Anyone considering the use of Xenazine must balance the risks of depression and suicidality with the clinical need for control of choreiform movements. Close observation of patients for the emergence or worsening of depression, suicidality, or unusual changes in behavior should accompany therapy. Patients, their caregivers, and families should be informed of the risk of depression and suicidality and should be instructed to report behaviors of concern promptly to the treating physician.

Particular caution should be exercised in treating patients with a history of depression or prior suicide attempts or ideation, which are increased in frequency in Huntington's disease. Xenazine is contraindicated in patients who are actively suicidal, and in patients with untreated or inadequately treated depression.

Xenazine is also contraindicated in patients who have impaired hepatic function or are taking monoamine oxidase inhibitors or reserpine. At least 20 days should elapse after stopping reserpine before starting Xenazine.

The need for therapy should be evaluated on an ongoing basis with the patient's doctor. Xenazine should be titrated slowly over several weeks for a dose that is appropriate for each patient. Before a dose greater than 50 mg is administered, the patient's CYP2D6 metabolizer status should be determined.

Neuroleptic malignant syndrome (NMS), akathisia, agitation, parkinsonism, dysphagia, and QT prolongation-related arrhythmias have been reported with use of Xenazine. Xenazine should not be used in combination with drugs known to prolong QTc (which in certain circumstances can lead to torsades de pointes and/or sudden death), in patients with congenital long QT syndrome, or in patients with a history of cardiac arrhythmias. A potentially irreversible syndrome of involuntary, dyskinetic movements called tardive dyskinesia (TD) may develop in patients treated with neuroleptic drugs. If signs and symptoms of TD appear in a patient treated with Xenazine, drug discontinuation should be considered. Adverse reactions associated with Xenazine, such as QTc prolongation, NMS, and extrapyramidal disorders, may be exaggerated by concomitant use of dopamine antagonists.

Xenazine elevates serum prolactin concentrations. Xenazine may induce sedation and somnolence (sleepiness or drowsiness) and may impair the ability to drive or operate dangerous machinery.

Some adverse events such as depression, fatigue, insomnia, sedation/somnolence, parkinsonism, and akathisia may be dose-dependent. If the adverse effect does not resolve or decrease, consideration should be given to lowering or discontinuing Xenazine. The most commonly reported adverse events with Xenazine compared to placebo were sedation/somnolence (31% vs 3%), fatigue (22% vs 13%), insomnia (22% vs 0%), depression (19% vs 0%), akathisia (19% vs 0%), anxiety (15% vs 3%), and nausea (13% vs 7%).

For more information, please see full [Prescribing Information, including Boxed Warning](#).

References

1. Rosenblatt A, Ranen NG, Nance MA, Paulsen JS. *A Physician's Guide to the Management of Huntington's Disease*. 2nd ed. New York: Huntington's Disease Society of America; 1999.
2. Haddad MS, Cummings JL. Huntington's disease. *Psychiatr Clin North Am*. 1997;20:791-807.
3. Huntington Study Group. Unified Huntington's Disease Rating Scale: reliability and consistency. *Mov Disord*. 1996;11:136-142.